

Nondysraphic cervicomedullary intramedullary lipoma

ABSTRACT

Spinal cord lipomas are usually associated with spinal dysraphism and is most common in lumbosacral region. Spinal intradural lipoma is a rare condition accounting for less than 1% of spinal cord tumours and is most prevalent in cervicodorsal region. Intramedullary spinal cord lipoma of cervical spine not associated with spinal dysraphism is one of the rarest lesions. They usually present insidiously with slowly progressive myelopathic deficits. We present a case of nondysraphic intramedullary spinal cord lipoma with exophytic component and intracranial extension. A 30 year male patient presented with wasting of muscles of left shoulder and left forearm with resultant weakness for last two months and myelopathic signs. Magnetic resonance imaging revealed a T1 hyperintense , T2 hypointense lesion on the dorsal aspect of cervical spinal cord with intracranial extension and exophytic component. There was no contrast enhancement, fat suppression image indicated a lipoma. The patient underwent cervical laminectomy with foramen magnum decompression with subtotal resection of intramedullary lipoma. Histopathology examination confirmed the diagnosis of lipoma. Subtotal resection of intramedullary lipoma is a reasonable and acceptable surgical option considering the benign nature of lesion and high probability of neurological deterioration due to dense adhesion between lipoma and neural tissue.

Keywords: Cervical cord tumor, intramedullary spinal tumor, lipoma, subtotal resection of lipoma

INTRODUCTION

Spinal intradural lipomas are rare lesions constituting <1% of all intramedullary spinal cord tumors.^[1] Most of the spinal cord lipoma are associated with spinal dysraphism and are usually located in the lumbosacral region. Intramedullary, lipoma not associated with spinal dysraphism is very rare.^[2,3] Most of them are localized in cervicodorsal region and present in the second and third decade of life.^[4]

CASE REPORT

A 30-year-old male patient presented with progressive weakness of shoulder and forearm muscles for 2 months. He also complained of numbness and paraesthesia of both upper limbs. There was evidence of progressive wasting of left shoulder muscles. Physical examination revealed significant wasting of left shoulder, left upper limb monoparesis, grade 3/5; proximal more than distal muscles, signs of cervical myelopathy in the forearm, and hyperreflexia. He did not have any gait disturbance or bowel bladder involvement.


Radiological workup revealed a dorsally located hyperintense lesion on T1-weighted image (T1WI) with cord compression [Figure 1]; which was hypointense on T2-weighted image (T2WI) at cervicomedullary junction extending upto C4 spinal segment [Figure 2]. Fat suppression images showed hypointensity [Figure 3]. There was no contrast enhancement [Figure 4]. There was no spinal dysraphism.

The patient was taken up for surgery in prone position with electrophysiological monitoring. Suboccipital craniotomy and posterior midline cervical laminectomy of C2–C4 vertebrae was done. A midline durotomy was performed and craniocaudal extent of tumor was delineated and exposed. Tumor

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Figure 1: T1-weighted image of cervicomedullary region (sagittal view) showing intramedullary hyperintense lesion with exophytic component and with intracranial extension



Figure 2: T2-weighted image of cervicomedullary lesion showing intramedullary hyperintense lesion



Figure 3: T1-weighted image with contrast showing insignificant contrast enhancement



Figure 4: Fat suppression images shows hypointense cervicomedullary lesion

was intramedullary in location with prominent exophytic component and was yellowish in colour. Subtotal resection of the lesion was done from dorsal to ventral direction under neurophysiological monitoring. Total excision was not possible due to dense adhesion between the lesion and ventral cord. Postoperatively, patient had some reduction in muscle power; patient showed neurological improvement with physiotherapy. Histopathology confirmed the diagnosis of lipoma.

DISCUSSION

Spinal cord lipoma account for <1% of all spinal cord tumors.^[1] Almost always they are located dorsal aspect of cord, subpial in location and hence are considered juxtamedullary.^[5] In most of the cases, they are associated with spinal dysraphism. Lipoma consists of mature adipocytes lying in fibrous connective tissue. Striated muscle, neural

tissue, epithelial tissue, and other mesenchymal derivatives may also be found in lipoma.^[6] The adipocytes do not have any atypical features.

Origin of lipoma

Intramedullary lipoma is the most common dysembryogenic lesions, accounting for 1% of all intramedullary tumor that has an embryological basis.^[5,7] It is contentious whether the lipomas are true neoplasm or hamartoma. Lipomas associated with spinal dysraphism are considered hamartomaous^[8] whereas nondysraphic lipomas are thought to be a true neoplasm.

Etiology of lipoma is not well established yet. Several hypotheses has been propounded to explain the intramedullary location of fatty tissue. The “developmental error hypothesis” postulates that mesenchymal cells, precursor to adipocytes become misplaced during neural tube development and migrate into the developing neural tube before closure, and

develop into a lipoma later.^[9] This explains dorsal location of the lipoma. Growth of lipoma displaces the dorsal roots more laterally. “Metaplasia theory” holds the metaplasia of connective tissue as the cause of formation of lipoma.^[10] The presence of peripheral nerves, dermoid cyst, skeletal muscle, and lymphoid tissue in intramedullary lipomas have been explained by hamartomatous theory.^[10,11] Another hypothesis proposes that adipocytes arise from the cells giving rise to spinal cells. In normal conditions, mesenchymal cells form spinal vessels but are capable of differentiating into adipocytes unless inhibited by neural crest cells. If neural crest cells are defective, the inhibition fails, and mesenchymal cells form adipocytes.^[12]

Imaging

Magnetic resonance imaging (MRI) remains the most sensitive modality for the tumor. Fat is hyperintense on T1WI, hypointense on T2WI with no contrast enhancement. Fat suppression sequence identifies lipoma from blood or calcification which remains hyperintense.

Pattern of presentation

Intramedullary, lipoma has two distinct patterns of presentation. One group manifest in early infancy and childhood as tetraplegia or floppy baby syndrome, probably due to birth trauma to the spinal cord.^[7,13]

The other group comprised adults, mostly in their second or third decade of life, who present with slowly progressive neurological deficit which may remain static for long durations. These lesions become symptomatic as enlargement of lesion occurs due to fat deposition in metabolically normal fat cells.^[5] In most cases, the clinical course is static. In some cases, they can progress slowly with neurological deterioration.

Surgical options

Management of spinal cord lipoma is controversial warranting a tailored approach which may include observation, biopsy, subtotal removal, gross total removal, and spinal cord decompression. Incidental lipoma can be managed by observation for a long period until there is neurological deterioration. For patients presenting with neurological deterioration, spinal cord decompression with subtotal resection may be the best option considering the dangers of neurological deterioration in attempting to remove an adherent tumor and the indolent course of the lesion on the other hand.^[14-16] Few cases of excellent result in total or near total removal have been reported.^[14]

Intraoperative ultrasound is very helpful delineate the extent of lesion as lipoma is anechoic.^[17] Lasers are very valuable

adjunct in excision of lipoma as the laser vaporize the fatty tissue due to their high water content. Needless to say, intraoperative neurophysiological monitoring improves postoperative outcome. Delayed instability rarely occurs following laminectomy for intramedullary tumor removal in adults. While laminoplasty may be a reasonable option, it is not required in the adult population.^[5]

Postsurgically, most of the patients have an uneventful course with variable improvement of neurological deficit. Intraspinal lipomas, being congenital lesions not only compress but would actually replace normal tissue during development resulting in less redundancy in the functional pathways. Hence, there is greater chance of permanent injury to the residual neural tissue postoperatively.^[3] Therefore, early surgical decompression is recommended before irreversible spinal cord dysfunction sets in, following which, surgical results are suboptimal.

CONCLUSION

Nondysraphic intramedullary lipoma of the spinal cord is one of the rarest lesions of congenital origin. Usually, they have an indolent course and can present in early childhood or early adult life depending on size and rate of growth of lesions. Patients have slowly progressive neurological deficit. MRI is the most sensitive imaging protocol. Due to the benign natural history of lesion, spinal decompression, or subtotal excision of lipoma seems to be the best bet against sudden neurological deterioration.

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Conflicts of interest

There are no conflicts of interest.

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